PostScript

LETTERS TO THE EDITOR

Inflammatory myofibroblastic tumour of appendix

Inflammatory myofibroblastic tumours (IMFTs) are rare, distinct clinicopathological entities characterised by a dense inflammatory cell component amid myofibroblastic proliferation. Although the lung is the site commonly associated, there are reports of extra pulmonary involvement. IMFTs of the gastrointestinal tract are extremely rare and there have been only three confirmed cases of involvement of the appendix. These benign tumours have distinctive clinicopathological features that distinguish them from inflammatory fibroid polyps, gastrointestinal stromal tumours, smooth muscle tumours, sclerosing mesenteritis, lymphoma and other malignant tumours.[1–6] This letter highlights the clinical and immunohistological features of an IMFT of the appendix.

A 34-year-old man presented with progressively worsening colicky pain in the right lower abdomen for the past 3 days, associated with low-grade fever and two episodes of vomiting. Clinical examination showed a tender right lower quadrant with a vague mass. Investigations were unremarkable except for a raised total white cell count and an increased erythrocyte sedimentation rate (12 800 cells/mm^3 and erythrocyte sedimentation rate 48 mm/h, respectively). Abdominal ultrasound showed features of an appendicular mass. Laparoscopy showed a diffusely inflamed, oedematous appendix adherent to the surrounding small bowel and covered by omentum. A rounded, well-delineated mass of about 6 cm in diameter, arising from the mid-third of the appendix along its antimesenteric border was also noticed. Mesenteric adenopathy was not obviously evident. The appendix, with the mass, was carefully separated from the surrounding intestine and an enbloc resection, including the mesoappendix, was carried out. The appendix measured 8×7×4 cm and the whole specimen weighed 40 g; the mass arising in the mid-third of the appendix measured 6×5 cm (fig 1). The appendix showed diffuse inflammation with the mid-third stretched over the mass. The appendix was slit longitudinally and no obvious breach in mucosal continuity was noted; inflammatory changes were seen within, and luminal obstruction at the level of the mass was evident. A cut section showed a greyish-pink smooth-surfaced mass with a focal area of necrosis surrounding a fecalith measuring 13 mm in diameter (fig 2). A small 8 mm lymph node was identified in the mesentery. Sections showed appendicular mucosa with ulceration, lymphoid hyperplasia and acute inflammatory cells. Sections from the mass showed spindle-shaped myofibroblasts amid inflammatory cells consisting of eosinophils, lymphocytes, plasma cells and neutrophils in order of frequency (fig 3). Blood vessels with irregular blood spaces of varying sizes were seen. No evident granulomatous changes (negative staining for acid-fast bacilli) or cellular atypia were observed. The lymph node showed features of reactive hyperplasia. Culture showed *Escherichia coli*. Immunohistochemistry showed intensely positive vimentin, smooth muscle actin staining in spindle cells and negative staining for neurone-specific enolase, S-100, desmin, CD34 and CD117. The patient's postoperative recovery was uneventful and he has no evidence of recurrence nearly 18 months after surgery.

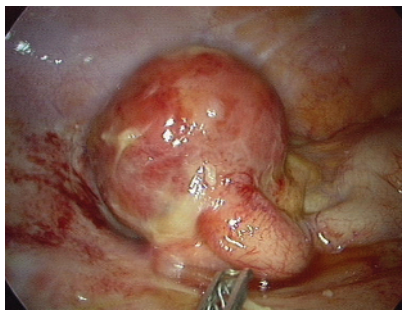

Figure 1 Gross appearance of appendicular mass (inflammatory myofibroblastic tumour).

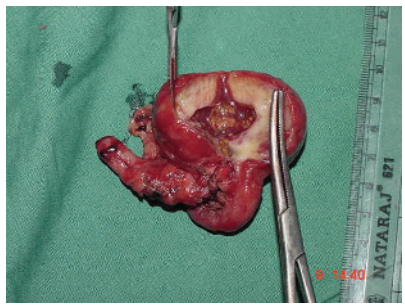

Figure 2 Cut section of appendicular mass showing abscess and fecalith in the centre.

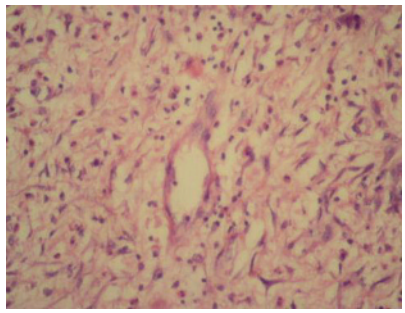

Figure 3 Inflammatory cells consisting of eosinophils, plasma cells, lymphocytes, neutrophils, histiocytes and spindle-shaped myofibroblasts (haematoxylin and eosin staining ×400).

Considered as an aberrant or exaggerated response of the tissues to a chronic inflammatory process, IMFT could be a histological expression of an infective or reparatory process and, occasionally, a true neoplasm.[2] The true incidence of these tumours is unknown; in addition, as immunostaining is not routine in most laboratories, many cases probably go undiagnosed and unreported. Generally singular, with no specific age or sex predilection,[3] their clinical manifestations usually relate to the organ associated, with no specific features for preoperative diagnosis. These are firm, solid, well-encapsulated tumours of varying sizes, with a gritty sensation on cutting and occasional areas of necrosis. Histologically, the tumours consist of dense infiltration, with inflammatory plasma cells, histiocytes, lymphocytes and eosinophils amid well-populated areas of spindle-shaped myofibroblasts. Three histological patterns are described: fibromyxoid or vascular, proliferating and sclerosing. Atypia is rare, with few or no mitoses.[3] The histological manifestations range from inflammatory cell proliferation to neoplastic spindle-cell formation, with various parts of the same lesion showing a wide range of processes. Varied aetiological factors include chronic irritating factors[5] and infections caused by *Helicobacter pylori, E coli*, Epstein–Barr virus, for example. In our case, the fecalith may have been a contributory factor that exaggerated the inflammatory response. Microabscesses or cavities with necrosis have been described; not all have yielded any detectable growth.[3 5] Immunostaining positivity for smooth-muscle-specific actin and vimentin is helpful in confirming the myofibroblastic origin, whereas reactivity to CD34 is negative. Anaplastic lymphoma kinase reactivity has been recently detected in some spindle-cell components of these tumours.[3]

Specific immunostaining techniques can help differentiate these tumours, with a good prognosis. Treatment constitutes complete excision with lymph node clearance, although the involvement of nodes is an exception rather than the rule. Recurrences and rare examples of malignant transformation have been reported.

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Glomangioma of the lungs: a rare differential diagnosis of a pulmonary tumour

Glomangiomas (glomus tumours) are uncommon, and in most cases are benign perivascular tumours, usually located in the dermis of the extremities, especially in the subungual region of the fingers. They also occur in other organs and in other regions of the body. Histogenetically, they are derived from modified smooth muscle cells of the glomus apparatus, which is associated with temperature regulation. These tumours are composed of capillaries, sometimes of dilated endothelial bigger vessels surrounded by nests of uniform, cytoplasm-rich cells with isomorphic round or ovoid nuclei (glomus cells). They also contain a variable amount of smooth muscle tissue. Three different types of glomus tumours are described according to their different components. The common form is predominantly made up of glomus cells, with a smaller amount of vessels and smooth muscle cells. The second variant, called ''glomangioma'', contains more partly dilated vessels resembling cavernous haemangiomas and is less circumscribed than the common form. In the most rare type, the ''glomangiomyoma'', the glomus cells undergo a transition to elongated smooth muscle cells.

Immunohistochemically, glomus tumours usually show a positivity for vimentin and smooth muscle actin and a negativity for cytokeratin, neuroendocrine markers (chromogranin and others), CD31, CD34 and S-100.[1]

These tumours are very rare in the lungs, and, to our knowledge, only 14 cases have been described, including two malignant glomus tumours.[2–10]

Case report

A 64-year-old man was admitted to the hospital for resection of a carcinoma of the rectum. The preoperative computed tomography showed a round tumour (3.5 cm) in the left lower lung lobe that was removed 2 weeks after the resection of the rectum.

Findings

On macroscopic examination, the round tumour was reddish. Histological examination (haematoxylin and eosin, Elastica-van-Gieson, Prussian Blue and periodic-acid Schiff) showed lung tissue with partly compressed, partly hyperaemic and ectatic vessels that exceeded the normal density of alveolar capillaries (fig 1A,B). Cytoplasm-rich cells with round isomorphic nuclei were arranged diffusely perivascularly and in nests. Immunohistologically, the tumour cells featured a cytoplasmic staining for α1-smooth muscle actin (fig 1A), whereas the reactions with antibodies against CD31, CD34, pancytokeratin and TTF-1 were negative.

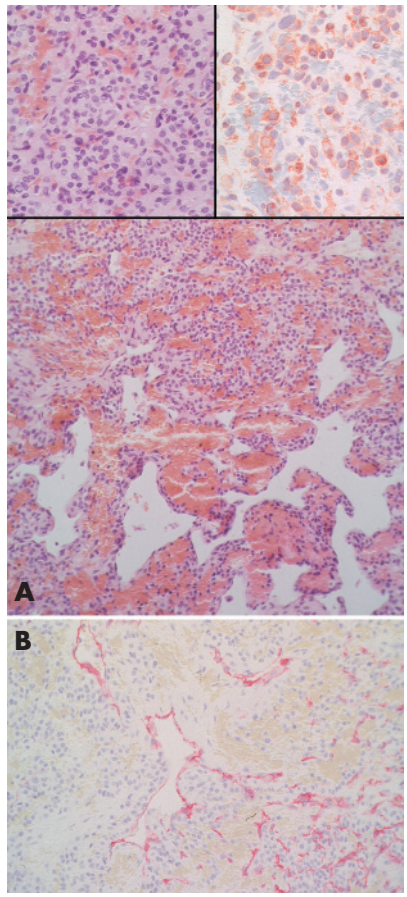

Figure 1 (A) Part of the glomangioma with many partly dilated vessels and the pericapillar cytoplasm-rich cells with small round isomorphic nuclei (haematoxylin and eosin; original magnification ×25). The left inset depicts the typical cells of the glomus tumour at a higher magnification (haematoxylin and eosin; original magnification ×100). The right inset shows cytoplasmic staining of the tumour cells with antibodies against α1-smooth muscle actin (original magnification ×100). (B) Transition of the glomangioma from the solid, cell-rich part (left) to the more vascularised area (right; CD34; original magnification ×50).

Comment

On the basis of the histological and immunohistological results, a metastasis of the rectal carcinoma was ruled out and the tumour was identified as a glomangioma of the lung.

The other important differential diagnoses, including carcinoid small-cell lung cancer, melanocytic neoplasms, haemangiopericytoma, smooth muscle neoplasms, paraganglioma and primitive neuroectodermal tumours, were also excluded by morphology and immunostaining pattern.

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Vertebral artery dissection revisited

Vertebral artery dissection is considered to be one of the most difficult dissections that a forensic pathologist has to undertake. Traditionally, vertebral arteries have been dissected using two time-tested procedures. The first procedure is carried out by removing the neck block and decalcifying the block.[1] The neck block can be removed using the anterior or the posterior approach. The anterior approach is less time consuming but is more difficult to carry out. The posterior approach, on the other hand, is more time consuming but is simpler to carry out. The other procedure requires the use of special instruments for dissection.[2] These are a Swann–Morton number 5 scalpel handle with number 11 blade, Swann–Morton number 3 scalpel handle with number 12 blade